

MD

Myotonic Dystrophy  
SUPPORT GROUP

Swallowing  
Difficulties  
in Myotonic  
Dystrophy

by  
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# Myotonic Dystrophy

Swallowing difficulties are an important aspect of Myotonic Dystrophy due to potentially serious complications. They should be identified early to help reduce risk of life-threatening complications. Management options often include swallowing strategies and sometimes alternative routes of feeding. These will be outlined as part of this booklet.

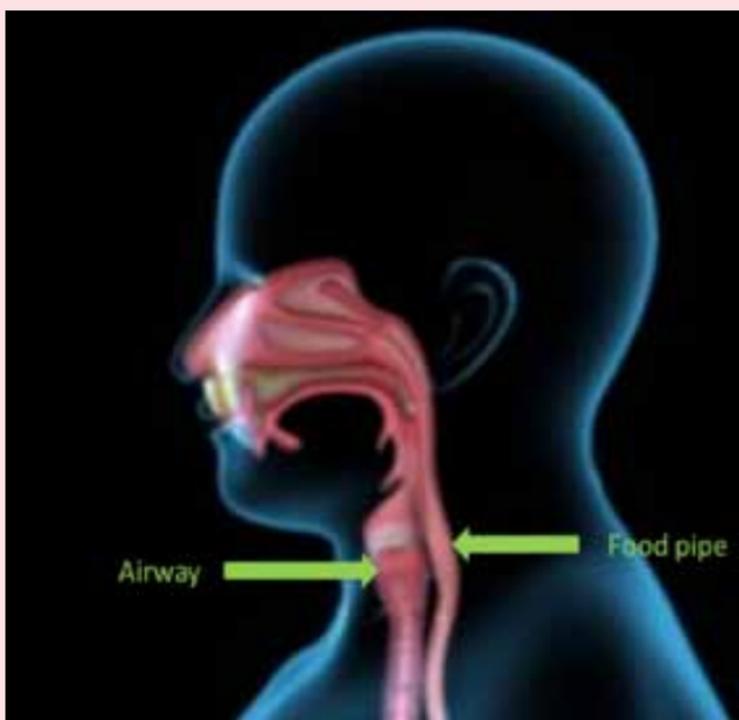
## **Brief Summary**

- Myotonic Dystrophy can affect the muscles of your face, mouth and throat.
- Weakness or stiffness (myotonia) in these muscles can cause problems with swallowing.
- Swallowing problems can lead to weight loss and chest infections.
- Problems can be identified early by looking out for signs and symptoms. These may include longer mealtimes, food sticking in the throat, needing to drink with meals and coughing and spluttering.

- Swallowing problems should be assessed by a Speech and Language Therapist who can provide you with tailored advice and options.

### **How do we normally eat and drink?**

The wind pipe (known as the trachea) and food pipe (known as the oesophagus) sit very close together in the throat, as shown below.



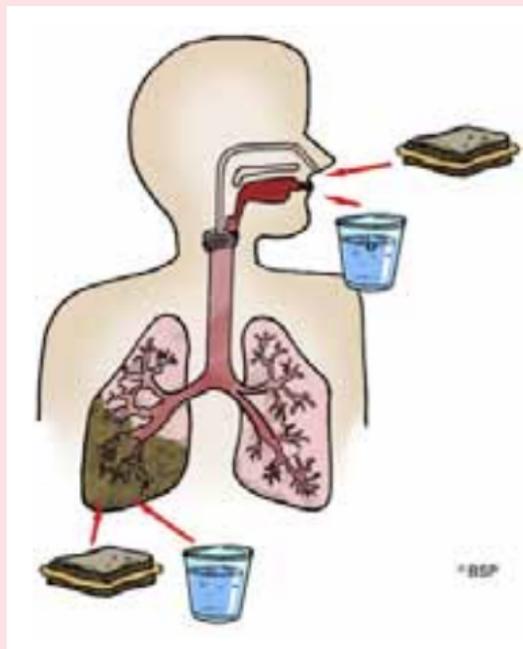
*The airway and food pipe sit close together in the throat*

When talking or engaging in an activity other than eating and drinking, the airway is open. This allows oxygen to pass into the lungs and expel waste gases out. At these times the entrance to the food pipe is closed.

At regular intervals we initiate 'a swallow.' This allows saliva to move from the mouth into the food pipe. At the point of swallowing, the airway closes, breathing is suspended and the food pipe opens momentarily. When the swallow is complete, the food pipe closes, the airway opens and breathing starts again. The same thing happens when we eat and drink. The swallowing mechanism is not 100% reliable. Most of time we can eat and drink without complication, however even with a 'normal' swallow, occasionally things can go wrong causing food, drink or saliva to 'go down the wrong way' causing coughing and spluttering. If the person coughing does not have an underlying disease, the cough is usually strong enough to move things out of the airway and no long-term complications occur.

## **What are swallowing difficulties and why do they occur?**

Problems with chewing and swallowing are known as 'dysphagia.' In Myotonic Dystrophy, changes usually occur as a result of weakness in the mouth and/or throat muscles. Weakness in the lips, tongue and throat can cause problems with chewing, clearing the mouth and clearing food and drink from the throat. In more severe cases food and drink can get stuck in the throat for quite some time and can 'go down the wrong way.' This means food and drink may enter the airway rather than the food pipe.



*Food and drink can go down the wrong way causing chest infections*

Food and drink in the airway is known as aspiration. Myotonia (an inability to relax muscle) in the jaw or tongue can also cause problems with chewing and swallowing in this disease.

### **Am I at risk of developing swallowing difficulties?**

Swallowing problems occur more frequently in some muscle diseases than others. Problems with swallowing are common in Myotonic Dystrophy but no one person is affected in the same way. Some people develop very few difficulties whilst for others it can be more problematic. Swallowing difficulties can change over time so even if you don't have any problems swallowing at the moment, you may develop some difficulties later on.

### **How will I know if I have changes in my swallowing?**

Changes in swallowing often occur very gradually, so gradually that you may not notice

them. Sometimes people make changes to the food they eat to make chewing and swallowing easier but don't realise they are doing so. Some of the things you may notice are:

- Taking skins off fruit
- Dunking biscuits rather than eating them dry
- Avoiding foods such as nuts, crisps or chewy meat
- Adding extra gravy to meals or taking crusts off toast

You may also notice things such as:

- Needing drinks with your meals to help 'wash' food down



*Small sips of fluid during a mealtime can help clear the throat*

- Mealtimes taking longer than before
- Needing to cough or clear your throat when eating
- Needing to swallow more than once to clear food or drink from your throat
- Episodes of coughing during or after meals



*Episodes of coughing or choking during or after eating are a sign of swallowing problems*

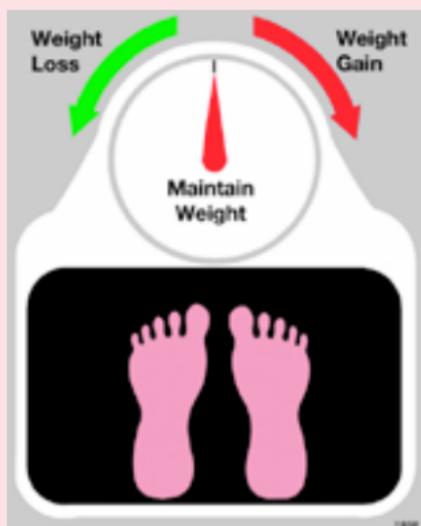
## **Why is it important to notice changes in my swallowing?**

It is helpful to notice changes in your swallowing early so that your doctor or therapy team can help minimise longer-term risks. It will also help you make informed and advanced decisions about your eating and drinking.

It is useful for you to know that longer-term problems with your swallowing can lead to:

- Weight loss
- Chest infections

*Dysphagia can contribute to unintentional weight loss*



The sooner we identify swallowing problems, the more we can do to prevent complications or consequences. It can be helpful to think back

to a memorable event a year or so ago to help work out whether things have changed with your eating and drinking. Difficulties with swallowing can be particularly problematic for people who already have problems with breathing and a weak cough. This is because they are less likely to be able to clear any food and drink residues from your throat or airway.

### **Will I always know if I have problems with my swallowing?**

Evidence suggests that people with Myotonic Dystrophy are not always fully aware of changes in their swallowing. It may be helpful ask a family member or carer whether they have observed any of the changes listed above.

### **What should I do if I am concerned about my swallowing?**

If you are concerned about your swallowing, you should request a referral to a Speech and Language Therapist via your GP, clinical nurse specialist or neurologist. If you are already known

to a Speech and Language Therapist, contact them again for an up to date assessment.

If you are really struggling with your swallowing and it is:

- Stopping you from eating and drinking altogether;
  - Causing persistent chest infections;
  - Or persistent coughing and choking events
- you should seek medical advice as soon as possible.

### **What can a Speech and Language Therapist do for me?**

A Speech and Language Therapist specialises in the assessment and management of swallowing difficulties. You may see them either in your own home, local clinic or hospital. In your first contact you should expect them to:

- Take a detailed history from you about your swallowing difficulties
- Assess your face, mouth and throat muscles
- Observe you eating and drinking

The Speech and Language Therapist should take into account factors such as your mobility, breathing difficulties and need for a ventilator before giving advice and recommendations about your swallowing. They may decide to refer you into a specialist clinic for an 'instrumental' assessment of your swallowing.

### **What is an instrumental assessment of swallowing?**

An instrumental assessment uses instruments to assess your swallowing in more detail. It allows the Speech and Language Therapist to visualise the inside of your mouth, throat and your airway. There are two possible assessments:

- A swallowing x-ray (known as a Videofluoroscopy, VFS)
- A mouth, throat and airway camera (known as a Fibreoptic Endoscopic Evaluation of Swallowing, FEES)

A VFS uses x-rays to see inside your mouth, throat and foodpipe. It is not invasive but does



*A videofluoroscopy (VFS) helps understand swallowing via x-rays*



*A Fibreoptic Endoscopic Evaluation of Swallowing (FEES) helps understand swallowing via camera.*

need to be done in a hospital with x-ray specialists. FEES uses a camera on the end of a small, flexible tube.



*A videofluoroscopy image highlights any issues with swallowing*

The camera is passed via your nose to visualise your throat. Neither assessment is painful. Your Speech and Language Therapist will provide you with more information and help you decide which assessment is best for you.

## **What is sensible advice to keep my swallowing as safe as possible?**

You will need an assessment from a Speech and Language Therapist to provide you with tailor-made advice. The following tips can help keep your swallowing as safe as possible whilst you are waiting for assessment. These are:

- Sit as upright as possible when eating and drinking
- Choose a quiet place to eat your meals. Don't talk whilst eating
- Take smaller mouthfuls
- Chew foods carefully
- Take your time, don't rush
- If you need assistance to eat and drink – make sure your carers know you need time to chew and swallow each mouthful. Go at your pace, not theirs
- Avoid or adapt foods you find difficult (for example cut crusts off bread, dunk your biscuits in tea or mince-down tough meats)
- Select 'easy' foods such as soft, easy-to-chew foods (for example fish rather than meat, cake rather than biscuits).

It is also a good idea to avoid eating whilst tired. If you tire during meals, consider having smaller meals more often rather than larger meals less often. If you feel food catching in your throat, try swallowing again or clearing your throat with a gentle cough. You may find it helpful to wash food down with small sips of drink; but don't gulp or drink quickly and wait until you have swallowed the food

If you use non-invasive mask ventilation (known as NIV), you should allow plenty of time after meals for food and drink to pass safely into your stomach before putting on your breathing mask. If you are new to using NIV, only eat and drink when the NIV is off. Over time some patients learn to control their breathing and swallowing on the NIV however this is not recommended, at least in the beginning.

If you do have problems with your swallowing, it is a good idea to have somebody around or easily contactable when you are having your meals in case you experience any difficulties.



## **What options are available to help with my swallowing?**

There are a small number of published research studies looking at the benefits of exercise, surgery and drug treatments for improving swallowing function in muscle disease. At the moment, there is no strong evidence to support these approaches in Myotonic Dystrophy. The research is however continuously evolving. Some therapists may provide you with exercises to try on an individual basis. They may also tailor individual advice to manage any jaw or tongue myotonia.

Most frequently, we aim to keep swallowing as safe and efficient as possible using swallowing strategies. These should be tailored to your individual needs. If we cannot maintain your weight or keep you chest infection-free using swallowing strategies, the Speech and Language Therapist (in discussion with your neurologist and dietitian) may talk to you about alternative ways to support your swallowing. This may include

non-oral feeding (also known as enteral feeding). This is feeding support that isn't via the mouth. Small feeding tubes can be placed into the stomach to by-pass the swallowing. An example tube is shown in the photograph below.



*A small feeding tube in the stomach can help support your swallowing*

Feeding tubes can help provide additional calories and reduce the risk of food and drink going down into the lungs. The benefits and risks of a feeding should be discussed on an individual basis. In general, they are well tolerated and can be used in addition to eating and drinking. The feeding tubes do not prevent you from taking food or drink by mouth and can be hidden under a blouse or shirt when not in use.

## **The Future**

Even if you don't have any problems with your swallowing, it is helpful for you to understand the potential for changes that may (or may not) occur over time. A good level of self-awareness and understanding can help us to manage any problems you may have proactively and hopefully minimise longer-term complications as much as possible.

## **Where can I get more information?**

The Myotonic Dystrophy Support Group may be able to help find other people affected by swallowing problems who you can talk to.

*We cannot accept responsibility for information provided by other organisations.*

## **Acknowledgements and References**

MDSG would like to thank:

- Heart of England NHS Foundation Trust
- Black Sheep Press Ltd © for allowing use of their images in this publication.

Further information about Myotonic Dystrophy can be obtained from:

- **Myotonic Dystrophy Support Group**  
19/21 Main Road, Gedling, Nottingham. NG4 3HQ  
Tel/Fax: 0115 987 5869  
Email: [contact@mdsguk.org](mailto:contact@mdsguk.org)  
[www.myotonicdystrophysupportgroup.org](http://www.myotonicdystrophysupportgroup.org)



## **Other publications available from the Myotonic Dystrophy Support Group:**

- Anaesthesia and Sedation for patients with Myotonic Dystrophy
- Basic Information for Midwives
- Bowel Problems in Myotonic Dystrophy
- Congenital Myotonic Dystrophy
- Excessive Daytime Sleepiness and Myotonic Dystrophy
- Facts for patients, family members and professionals
- Myotonic Dystrophy and the Brain
- Myotonic Dystrophy and the Eye
- Myotonic Dystrophy Support Group
- Relatives Information
- The Heart and Myotonic Dystrophy
- Why do we get new families with Myotonic Dystrophy?



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Myotonic Dystrophy Support Group Helpline 0115 987 0080





*Myotonic Dystrophy*  
SUPPORT GROUP

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